separated by 374 basepairs. *ABCG5* and *ABCG8* are both encoded by 13 exons and each spans ~28 kb. (B) The mutations detected in patients with sitosterolemia (Table 2) are indicated on a schematic model of *ABCG5* (left) and *ABCG8* (right) (C) Predicted amino acid sequence of ABCG5 and ABCG8, which are 651 and 673 residues in length, respectively. Alignment of the inferred amino acid sequences indicates 28% sequence identity and 61% sequence similarity between *ABCG5* and *ABCG8*. Both proteins are predicted to contain six transmembrane segments using the program MEMSAT 2 (Jones, *et al.*, *Biochem.* 33:3038 (1994)). The putative transmembrane segments of each protein are indicated by cylinders (B) and lines (C). The Walker A motif and Walker B motifs are highlighted. The ABC signature sequence (C-motif) is indicated.--

Q1

Please replace paragraph **[26]** beginning at page 8, line 8, with the following:

--**[26] FIGURE 3.** (A) ABCG8 exon 2 (reverse strand) through ABCG5 exon 2 (forward strand) (SEQ ID NO:9). The four exons are underlined and the conserved regions are in uppercase. The sequence ends in intron 2 of ABCG5 and is in the following order: ABCG8--exon 2 (reverse strand); ABCG8--intron 1 (reverse strand); ABCG8--exon 1 (reverse strand); gap between genes; ABCG5--exon 1 (forward strand); ABCG5--intron 1 (forward strand); ABCG5--exon 2 (forward strand); and ABCG5--intron 2 (forward strand, partial). (B) The sequence between ABCG5 and ABCG8 in which the control sequences (*e.g.*, bidirectional promoter, *etc.*) reside (SEQ ID NO:10).--

Q2

Please replace paragraph **[117]** beginning at page 33, line 20, with the following:

--**[117]** The particular expression vector used to transport the genetic information into the cell is not particularly critical. Any of the conventional vectors used for expression in eukaryotic or prokaryotic cells may be used. Standard bacterial expression vectors include plasmids such as pBR322 based plasmids, pSKF, pET23D, and fusion expression systems such as GST and LacZ. Epitope tags can also be added to

Q3

recombinant proteins to provide convenient methods of isolation, e.g., c-myc, HA-tag, 6-His (SEQ ID NO:11) tag, maltose binding protein, VSV-G tag, anti-DYKDDDDK (SEQ ID NO:12) tag, or any such tag, a large number of which are well known to those of skill in the art.--

Please replace paragraph **[198]** beginning at page 56, line 15, with the following:

--**[198]** Common linkers such as peptides, polyethers, and the like can also serve as tags, and include polypeptide sequences, such as poly-Gly poly-gly sequences of between about 5 and 200 amino acids (SEQ ID NO:13). Such flexible linkers are known to persons of skill in the art. For example, poly(ethelyne glycol) linkers are available from Shearwater Polymers, Inc. Huntsville, Alabama. These linkers optionally have amide linkages, sulfhydryl linkages, or heterofunctional linkages.--

Please insert the accompanying paper copy of the Sequence Listing, page numbers 1 to 23, at the end of the application.

REMARKS

Applicants request entry of this amendment in adherence with 37 C.F.R. §§1.821 to 1.825. This amendment is accompanied by a floppy disk containing the above named sequences, SEQ ID NOS:1-13, in computer readable form, and a paper copy of the sequence information which has been printed from the floppy disk.

The information contained in the computer readable disk was prepared through the use of the software program "PatentIn" and is identical to that of the paper copy. This amendment contains no new matter.

Attached hereto is a marked-up version of the changes made to the Specification by the current Amendment. The attached pages are captioned **"VERSION WITH MARKINGS TO SHOW CHANGES MADE."**